ON THE SO-CALLED FAMILY OR HEREDITARY FORM OF LOCOMOTOR ATAXIA.*

By WILLIAM A. HAMMOND, M.D.,

PROFESSOR OF DISEASES OF THE MIND AND NERVOUS SYSTEM IN THE NEW YORK POST-GRADUATE MEDICAL SCHOOL; SURGEON-GENERAL U. S. ARMY (RETIRED LIST); PRESIDENT OF THE AMERICAN NEUROLOGICAL ASSOCIATION, ETC.

TWENTY years ago, when doubt existed relative to the seat and nature of the lesion constituting the disease now generally known as locomotor ataxia, Friedreich,[1] of Heidelberg, published an elaborate essay, of which the title, "On degenerative atrophy of the posterior columns of the spinal cord," indicated the view which he entertained of its morbid anatomy.

The monograph was based upon six cases, and of these three died before the publication of the paper, and were submitted to very thorough *post-mortem* examination.

The first and second cases were brother and sister; the third, fourth, and fifth cases were sisters, and the sixth case was brother to these. The six cases were therefore comprised in two families. As they have been referred to by almost every writer on the subject since Friedreich, as constituting a peculiar form of locomotor ataxia, I think it desirable to give a few details of their symptoms, and of the results of the *post-mortem* examinations, before proceeding to the description of my own cases. I do this with the less hesitation, for the reason that all writers on the subject, so far as my knowledge extends, have accepted, without question, Friedreich's view of their character, while no one has thought it worth while to study them in the light of our present knowledge.

---

* Read before the American Neurological Association, June 23, 1882.

[1] Ueber degenerative Atrophie der spinalen Hinterstränge. *Virchow's Archiv*, B. xxvi, pp. 391 and 433; and B. xxvii, p. 1.

CASE I.—Andreas Lotsch, 33 years of age, was attacked with gradually increasing weakness of the lower extremities, so that walking became difficult. At the end of a year the paralysis was so well marked that the patient was unable to stand. During his twenty-sixth year the arms, first the right and then the left, became similarly affected, and a year later the speech began to be difficult and indistinct. There were no headache, vertigo, or pains, or other disturbances of sensibility in the limbs. The mind remained unaffected. Sleep, digestion, respiration, etc., were natural. For several years, however, erections had not occurred.

When the patient came under Friedreich's observation he was not able to stand, and had been in this condition for fourteen years. There was no atrophy of any of the muscles. The speech was so indistinct that he could hardly be understood. The sensibility of the skin in all parts of the body was not diminished in the slightest degree, the patient being able to feel the least prick of a pin, or the gentlest brush of a feather. He could, with the eyes closed, readily determine the differences in the weights of bodies, the position of his limbs, and their distance from each other. The electric contractility of the muscles was markedly diminished. The hearing, smell, taste, and sight were unimpaired. The pupils were equal, and reacted readily to light ; the mental faculties were unaffected. Appetite and urinary and intestinal functions perfect.

At the end of 1859 gastric symptoms and convulsive movements of the extremities occurred. The weakness increased, the brain became involved, and on the 8th of January, 1860, the patient died in a state of deep stupor.

The *post-mortem* examination showed the existence of intense hyperæmia of the dura mater, the pia mater, and the white substance of the brain. The membranes of the cord were also congested. The cerebro-spinal fluid was increased in quantity. The white substance of the cord appeared to be softened throughout its entire thickness, but the gray matter presented no obvious abnormal condition.

Microscopically, it is stated that the nerve fibres of the posterior columns were atrophied, and that in the place of the nerve elements there was a deposit of minute fibres, together with a minutely nucleated mass of granular substance and many corpora amylacea. The lateral and anterior columns were normal, as were also the gray substance, "there being no pathological change discovered by microscopical examination." The posterior roots of the nerves were atrophied, while the anterior roots, neither macroscopically nor microscopically, exhibited any deviation from the normal standard.

Now it requires very little consideration of this case to satisfy us that it was not one of locomotor ataxia. It scarcely presents a single symptom of that disease. As to the microscopical examination, it is almost valueless, owing to the imperfection of the methods for preparation then employed, not only by Friedreich, but all other observers, and which were such as to absolutely prevent the detection of the morbid conditions characteristic of the several spinal-cord diseases.

CASE 2.—Charlotte Lotsch, a sister of the previously described patient, had suffered from chlorosis from her fourteenth to her seventeenth year; during her eighteenth year she was first affected with a well-marked and permanent weakness, first in the left, and subsequently in the right lower extremity, of so severe a form, that it was with the utmost difficulty she could attend to her household duties. At night she had pains in her limbs. Up to this period the upper extremities and the speech were unaffected. At about her twenty-sixth year the upper extremities began to show weakness, and the speech to become indistinct. In July, 1858, in her thirty-first year, the disease having then lasted thirteen years, she came under Friedreich's care.

At this time her body was well nourished. She could not, however, walk, and standing, though possible, was attended with much difficulty, "because of the weakness in her knees." As she lay in the bed she could stretch and extend her legs with ease, and was able to direct the movements of the arms and hands to the performance of actions not requiring much strength. Continued movements were, however, difficult. If, for instance, she attempted to pick up a needle, the inco-ordination of the muscles was at once perceived, and it was only after repeated trials that she succeeded. Her speech was indistinct, but she could still be understood. Fibrillary contractions were noticed in the tongue. The cutaneous sensibility was absolutely normal. Electric excitability of the muscles was intact. Senses undisturbed, as was also the intellect. The pupils were equal, and responded well to light. Sleep, appetite, excretion of urine normal. Bowels somewhat constipated.

A year afterward she had a sudden attack of hemiplegia, affecting the right arm and leg, and a sensation of numbness in these parts.

Subsequently the symptoms all seemed to come to a standstill, and the patient was living at the time of the report of the case.

No one, I think, at the present day would consider this a case of locomotor ataxia.

CASE 3.—Justine Süss was healthy till her sixteenth year, when she experienced in her left leg a sense of weariness and weakness, which subsequently affected the right leg also. At the same time she suffered from tearing and darting pains in both lower extremities, which, when she appeared at the hospital, were only occasionally felt. About the twentieth year the right arm, and then the left arm, became weak, while the paralysis of the lower extremities had greatly increased. During the years 1857 and 1858, she had pains in the ends of the fingers, especially those of the right hand. Contractions of the legs and arms also ensued, to such an extent that, while lying down, the legs were often forcibly drawn up against the abdomen. The speech also became stammering and indistinct. There were frequent attacks of headache and vertigo.

On entering the hospital under Friedreich's care she was found to be well nourished, muscles well developed, appetite good, bowels somewhat constipated, urine natural, sleep generally good, though liable to be disturbed by the pains in the left lower extremity; mind good; nystagmus in both eyes. Pupils normal. Speech bad. Standing without support was impossible. Examination of the skin with the electrical brush showed the sensibility to be undiminished. She could also readily perceive the slightest prick with a needle or touch with the finger. Testing with the æsthesiometer gave normal results so far as the upper extremities, the front of the chest, and the face were concerned, but her answers as regarded the abdomen, the back, and the lower extremities, were so contradictory as to lead to the belief that a certain amount of numbness existed in these parts. Three months after her admission she died with a complication of head and abdominal symptoms.

A very minute *post-mortem* examination was made, the details of which I omit, except so far as regards the spinal cord. Here it is stated that there was inflammation of the membranes and degeneration of the posterior columns, the appearances being similar to those observed in Case 1. Nothing is said of the anterior or lateral columns, or of the gray substance.

There is nothing in this case to show that it was one of locomotor ataxia. The pains complained of as being sometimes present in the upper and lower extremities were doubtless due to the implication of the lateral columns in the morbid process.

CASE 4.—Salome Süss, younger sister of the foregoing patient, was affected in her fourteenth year with lateral curvature of the spine and palpitation of the heart. At about her sixteenth or seventeenth year she suffered from weakness of both lower extremities, with rending pains. The paralysis of the lower limbs increased, and by her twentieth year had reached the upper extremities, but without pain. At her twenty-sixth year the speech began to be difficult. Cramps, mainly in the peroneal muscles, appeared. Excretion of urine and fæces normal.

When first seen by Friedreich, the speech was bad; the arms could be moved tolerably well, but without precision. Buttoning the clothes, picking up a needle, etc., were no longer possible. The pressure of the hands was slight; while lying in bed the patient could move the legs in all directions, but was unable to walk or stand alone; when in a sitting posture "she fell together," as it were. Sensibility of the muscles and electric contractility normal. The cutaneous sensibility in all parts of the body, as shown by the æsthesiometer, was perfect, and the least touch was at once felt. Senses good. The pupils were moderately large, and reacted well to light, but when the eyes were directed to a distant object the right pupil was larger than the left.

Soon after entering the hospital, head and abdominal symptoms were developed, and in the course of a couple of weeks she died.

The membranes of the cord showed evidences of inflammation. The posterior columns were found to be atrophied. The anterior and lateral columns were normal. Microscopically, the results were similar to those previously obtained, except that the lateral columns were also involved. The anterior columns were normal. The gray substance is not mentioned.

CASE 5.—Lisette Süss, eldest sister, being thirty-six years old, was healthy till her fifteenth year, when she began to experience continuous weakness and fatigue in both lower extremities, which gradually extended to the upper extremities also. "Growing pains in the bones," as she expressed it, were often felt. The weakness, little by little, advanced, and eventually the speech became involved.

When Friedreich first saw her she was confined to her bed, being unable either to walk or stand, and had suffered with her disease about twenty years. She could move her legs in all directions while lying down, but they doubled up under her as soon as she attempted to stand. The speech was so bad that she could hardly be understood, and in attempting to articulate long words she was obliged to break off and begin again more than once. There was no paralysis or other trouble with the tongue, so far as its appearance went. The cutaneous and muscular sensibility were normal. Mind unimpaired. Pupils of medium size, equal, and of normal reaction to light. When attempting to sit up she "went together." From an early period of her disease the patient had exhibited a tendency to nervous attacks of a hysterical character. These were evidently hystero-epileptic, as she was convulsed, uttered cries, and threw herself about the bed. After one of these seizures she would complain of neuralgic pains in the lower extremities. She was living at the time of the report.

This case does not differ essentially from the last two. I fail to discover any evidence of locomotor ataxia.

CASE 6.—Friedrich Süss, twenty-four years old, brother of the three patients whose cases have just been cited. As a child, was healthy; during his fifteenth year suffered from a weakness of the left leg, which, about a year and a half or two years subsequently, appeared in the corresponding arm. Soon afterward the right leg and right arm were similarly involved, so that when he was seventeen years old he could not use any of his limbs with their ordinary strength; during his eighteenth year the speech became difficult. For about a year he had experienced slight formication in the left arm.

When seen by Friedreich he could walk without assistance but with difficulty and insecurely. He could give a pretty strong grasp with the hand, and the action of individual muscles was good but when movements requiring a certain degree of co-ordination and delicacy were attempted trouble was experienced. For instance, the picking up of a needle was only accomplished after repeated trials. The speech was bad, the patient being understood with difficulty. Examination with the æsthesiometer showed the tactile sensibility to be normal in all parts of the body. Mind unaffected. Pupils were of moderate size, equal, and reacted normally. No derangement of the functions of the bladder or rectum. Sleep good. Three years afterward the disease was found to have made considerable progress, but no new symptoms had been developed. Standing and walking with the eyes shut, or in the dark, were difficult, but individual muscles contracted well through the influence of the will. The sensibility of the skin, the electric contractility of the muscles, the smell, taste, sight, and hearing were unimpaired. No changes in the pupils. Mind good; no headache, but at times slight vertigo.

As I have already said, no neurologist of the present day, after a full consideration of these cases, would regard them as instances of locomotor ataxia. That they have been so regarded by competent authorities[1] can only be explained upon the hypothesis that they have not been thoroughly studied, and that one writer has accepted the statements of another without referring to the original source.

Thus we find that in every case the first symptom observed was weakness of the lower extremities, and that this gradually extended so as to involve the upper extremities. Here was, therefore, a true paralysis.

---

[1] Among others, ERB, art. "Tabes Dorsalis," *Ziemssen's Handbuch*, elften Band, p. 194. LEYDEN, art. "Die Sclerose der hintern Rückenmarksstränge," *Clinic der Rückensmarks.-Krankheiten*, p. 324 *et seq.* ROSENTHAL, *Klinik der Nervenkrankheiten*, p. 363. ROSS, "A Treatise on the Diseases of the Nervous System," pt. ii, p. 235. BRAMWELL, "The Diseases of the Spinal Cord," p. 222.

We find also that in no single case, at any period of its course, was there the slightest loss of cutaneous or muscular sensibility.

In no case was there any derangement of the excretion of urine.

Without going any further I have no hesitation in saying that a case exhibiting these phenomena cannot be one of locomotor ataxia.

In addition, when the lancinating pains are not invariably met with—in fact, being very rare, when there is no swaying of the body on closure of the eyes, when the inevitable result is that the morbid process extends upward, that the speech becomes affected, that the pupils are always equal, not contracted, and reacting perfectly to light, I think we have an *ensemble* of symptoms which are absolutely incompatible with the idea of primary degeneration in any part of the posterior columns of the cord.

In a subsequent paper Friedreich[1] again alludes to the cases I have cited, and brings forward three others presenting similar features, and occuring in three sisters. And, again,[2] he gives the details of the *post-mortem* examination of one of the patients who had died of typhus. The principal features revealed were the facts that the posterior columns of the cord (the columns of Burdach and the columns of Goll) were in a state of sclerosis, and that the like condition existed in the medulla oblongata.

A case of a woman is detailed by Carre,[3] in whom all the prominent symptoms met with by Friedreich were present, and of whom it is stated that eight members (seven of whom were brothers and sisters) of the patient's family were similarly affected. She could walk equally well with the eyes closed as with them open, and did not have to look at her feet while walking.

---

[1] Ueber Ataxie mit besonderer Berücksichterung der hereditaren Formen." *Virchow's Archiv*, B. 68, 1876, p. 145.

[2] *Virchow's Archiv*, B. 70, 1877, p. 140.

[3] "Nouvelles Recherches sur l'Ataxie locomotrice progressive," Paris, 1865, pp. 65 and 248.

With these introductory remarks, I come to the consideration of the instances similar to those reported by Friedreich, which have come under my own observation.

CASE 1.—J. T., aged 9, was brought to me by his father, April 22, 1866. When four years old it was observed that he often fell even when walking or running on a level surface. In a year, about, he was unable to stand without support, and in the course of another year, his arms were noticed to be similarly affected. His speech which had been good up to this time, now became drawling and hesitating. No very rapid advance took place for about two years, and then the legs began to get still weaker, so that when he attempted to stand they doubled up under him. Soon after this he was brought to me.

On examination, I found paralysis more or less complete of all four limbs. They were smaller than was natural, but there was no atrophic degeneration. When lying down, the legs could be extended, flexed, abducted, and adducted. Tickling the soles of the feet at once produced involuntary withdrawing of these members. The sensibility of the skin was unimpaired. The prick of a pin was felt at once, and the sensation was invariably referred to the right place. There was no paralysis of the bladder or of its sphincter. With the eyes closed, the patient could put the index finger of either hand on any part of the body that might be designated. The speech was so indistinct that he could be understood with difficulty. The pupils were large, equal, and of normal reaction. All the special senses were intact. Mind good.

CASE 2.—H. T., aged 11, brother of the preceding patient, was brought at the same time. About six months previously the legs became gradually weak. At present there is no other noticeable symptoms. Can stand with the eyes shut and walk as well with them closed as open. Feels great weakness, so that after walking a hundred yards or so, is obliged to sit down. Gait staggering, but not at all resembling that of locomotor ataxia.

These patients were still living in 1876. The first was unable to stand or even to sit. The second could still stand alone, but could not walk without assistance. In the second the progress of the disease has been much less rapid than in the first, though the arms were markedly affected, and the speech was indistinct. No hereditary tendency is known to exist in the family.

CASE 3.—M. B., male, aged 10, was sent to me by Dr. Hubbard, of Bridgeport, May 11, 1872. When about six years of age he began to stumble and fall, and to complain of fatigue after very slight exertion in walking. The weakness in the lower extremities gradually extended to the arms, and also to the muscles which support the back and head. After two years the speech became affected, so that it was drawling, hesitating, and indistinct.

When he came under my notice he was unable to walk without assistance, though he could still stand alone. His attitude was that of an old man, the body being bent forward. In sitting, after a few minutes he "fell together," his body bending forward and the head settling down on the chest. It was impossible for him to feed himself, the muscles of the upper extremities being too weak to enable him to handle a knife and fork or to convey food to his mouth. There was also, at times, choking from difficulty of swallowing. The speech was so bad that no one but his mother could understand him. He had at no time suffered pain, and he was uniformly good-tempered and cheerful. The cutaneous and muscular sensibility were entirely normal, and the body was well nourished. The pupils were equal, rather large than otherwise, and reacting perfectly to light. There were no head symptoms, but the mind had evidently suffered from arrest of development. The functions of the bowels and bladder were undisturbed. The general health was good. There were frequent attacks of vertigo.

In walking he was obliged to use a cane or to hold some one's hand. Could walk as well with the eyes shut as when open, and there was no swaying of the body when the eyes were closed. The gait was not that of an ataxic, but rather of one suffering from simple motor paralysis.

This patient remained under my observation almost up to the time of his death, at the age of fourteen. I did not see him, however, when that event took place, but have been informed by his mother and by Dr. Hubbard that for the last four months of his life he was confined to bed, and that he suffered greatly from muscular spasms and contractions, the legs being frequently drawn up against the chest and held there forcibly for several hours at a time. At these periods there were severe pains in the head, spine, and in the limbs.

CASE 4.—C. B., brother of the preceding patient, and two years younger, was in good health till he attained his eighth year. He then began to complain of weakness in the legs, and in the course of a year was unable to walk without assistance. The arms during that period became similarly affected, and the case went on with symptoms nearly identical with those of his brother, till he died at the age of fifteen. At no time till shortly before his death were there any pains, and the cutaneous sensibility remained unimpaired through the whole course of the disease, as did also the functions of the bladder and bowels. The father and mother of these boys are of excellent constitutions, and there is no hereditary tendency to any affection of the nervous system, so far as can be ascertained. There was one other child who was accidentally drowned while an infant.

CASE 5.—E. M., thirty years old, came under my care when sixteen years of age, and has been seen by me at intervals since that time. The disease began when he was three years old, with weakness affecting first the right lower extremity, and gradually extending to the left. In the course of three or four years the upper extremities became affected in a similar manner, and at about the same time it was observed that the speech was beginning to be indistinct and drawling.

When I first saw this patient he could walk without assistance, though his gait was staggering. He could stand with the eyes shut, but his body swayed a little. His cutaneous sensibility was perfect, as were also all his other special senses. Mind clear; pupils equal, somewhat dilated, and reacting normally. No well-marked nystagmus, though occasionally, as I was told, there were slight twitchings of the eyeballs. No head symptoms except vertigo; bladder normal; bowels at times constipated. When lying down he could move the legs in all directions, though with greatly diminished force. When standing and supporting himself with his hands he could place either foot with precision and deliberateness on any spot indicated. He could also direct the hands with exactness, but the grasp of the right on the dynamometer was only 60°, and of the left 45°. Had trouble in buttoning or unbuttoning his clothes, but more, I was satisfied, from muscular weakness than from incoördination. Has never had pains of any kind. At the present time the patient though worse is not greatly so. His mind is clear; there is slight difficulty in pronouncing words, and he often forgets them; has pains in the back of his head and neck, though in no other part of the body. Special senses all good, except his eyesight, which is bad at night, and after looking steadily at an object for a short time it appears blurred. Nystagmus. His muscular power is greatly impaired, though the electric contractility of all his muscles is good. Staggers when he walks. In moving about the room holds on to objects, and in the street uses a cane. Cannot stand with the eyes shut. If he falls it is always backward. Bladder and bowels normal. Tendon reflexes all abolished. Sometimes has involuntary twitchings of the hands. Is gradually but very slowly getting worse.

CASE 6.—H. M., brother of the preceding patient, is at the present time twenty-six years old. At seven years of age weakness of both lower extremities was observed. This patient was never under my professional care. Though I have recently subjected him to thorough examination, I cannot find any essential points of difference between his symptoms and those of his brother, except that they are not so pronounced. He can still walk without support, and co-ordinates tolerably well, better with the legs than with the arms. Has never had pains of a decided character, though sometimes, as he says, he "aches all over." Is deaf in left ear, from measles. Patellar tendon reflex abolished. Thinks he is getting better.

There is no hereditary tendency in these men to any neurosis, so far as can be ascertained. The father is a strong, well-made man, engaged in a large manufacturing business.

The following cases were reported to me by Dr. E. G. Coleman, of Hollywood, Arkansas.

CASE 7.—G. R. was attacked with paralysis of a general character about the period of the first dentition, which increased to the time of his death at five years of age. He could neither walk nor use his arms, and his speech was affected. His limbs retained their rotundity to the end. Sensibility always good; no pains.

CASE 8.—L. R., brother. Paralysis was first observed when he was six months old, and gradually increased. Case similar in all respects to the preceding. Died when three years old.

CASE 9.—W. R., brother. Impairment of motion was observed at an early period. Arms and legs retained their plumpness till death, which took place at three years of age. Speech affected.

A daughter remains perfectly healthy.

The following cases are reported to me by Dr. W. C. Warren, of Holly Springs, Mississippi.

CASE 10.—Girl now nineteen years of age. Disease began, when about nine years old, with weakness in legs, and a tottering and unsteady gait. Symptoms worse in the morning on rising from bed. Gradually they grew worse, until at last she was unable to walk a step. Then the upper extremities began to be affected so that she could scarcely extend the arm to shake hands with any one. Total want of co-ordination now exists in upper extremities, and the mind is getting weaker. Difficulty of articulation has been present for a year. No pain anywhere except, of late, below the knees. When sitting with the feet pendent and not touching the floor, the toes drop far below the heels. When lying in bed she assumes the form of a hoop, the knees being drawn up to the chin and the spine curved forward.

CASE 11.—At about six years of age her young brother began to exhibit symptoms of a similar character, and for four years has continued to walk with more and more difficulty. No pains.

CASE 12.—About a year or fifteen months since, a brother has become diseased in a similar manner. No pains.

All these children were healthy up to the time of the appearance of the paralysis. Their grandmother has been paraplegic for twenty years. No other nervous affection in any member of the family. All three of these children, the doctor states, are restless at night, frequently crying out, and starting from their sleep. Both the boys can walk with the eyes closed, and their minds are still good, and speech is as yet unimpaired.

I have already given some of the reasons which prevent me regarding the cases reported by Friedreich and Carre as being instances of locomotor ataxia. Those which I have brought forward were similar in all essential details, and are, I think, to be embraced in the same category. To state the matter more fully:

We have here a total of twenty-two cases of almost identical morbid physiognomy—the disease beginning with weakness in the lower extremities, and then gradually advancing upward with nearly complete freedom from pains, and in none of which were there the peculiar sharp, lancinating, electric-like pains met with in locomotor ataxia.

In all the cases the pupils, where any mention is made of their condition, were equal and of normal size and reaction.

In locomotor ataxia it is the rarest phenomenon to find the pupils unaffected when the lesion is situated in the upper region of the cord. I cannot recall, in the whole range of my experience, a single case of the kind in question in which there was not some aberration from their normal state or functions.

In all these cases the speech was involved in a peculiar manner.

In locomotor ataxia speech-derangement is exceedingly rare. I have never seen a case in which it existed.

In no one of these cases was there the slightest loss of cutaneous sensibility.

I believe locomotor ataxia to be impossible without anæsthesia at some time or other of its course. In no case was there any derangement of the functions of the bladder.

Only one case of the advanced disease in which such symptoms were absent has ever come under my observation.[1] In locomotor ataxia there is almost invariably impairment of the contractile power of the bladder, or of its sphincter, or of both.

---

[1] "Clinical Lectures on Diseases of the Nervous System," New York, 1874, p. 141.

In no case was the feeling of constriction around the body experienced.

As is well known this is a common symptom of locomotor ataxia.

In all the cases there was a gradual advance of the disease upward, so far at least as the symptoms were concerned. Such a uniform progression toward a common end is not witnessed in locomotor ataxia.

The gait of the persons affected who came under my observation was altogether different from that of ataxics. Instead of the feet being put down with a jerk and in two distinct movements, they were moved exactly like those of a drunken man when he attempts to walk.

The fact of there having been well-marked incoördination in several of the cases is not in itself, in the slightest degree, pathognomonic of locomotor ataxia; it being well known that incoördination is met with not only in other spinal affections, but in certain diseases of the brain. Finally, locomotor ataxia is very rare with children, whereas all the cases described began in persons in an adult age.

For these reasons I cannot consider the cases in question to be instances of locomotor ataxia or sclerosis of the columns of Burdach. Doubtless, in some of them these columns were involved to some extent, but it was in all such clearly a secondary phenomenon.

Friedreich lays considerable stress upon the alleged tendency of the disease to affect the female sex from the fact that of his nine cases all were women but two. On the other hand, all six of my cases were in males; the three reported by Dr. Coleman were also males, as were likewise two of the three reported by Dr. Warren. Of the twelve American cases, therefore, eleven occurred in males, and of the whole twenty-two cases referred to in this memoir, thirteen were males and nine were females.

In the absence of sufficient *post-mortem* evidence I hesitate to assign a locality to the lesion which constitutes the pathological entity of the cases to which I have referred. I am inclined to think, however, that its primary seat is the medulla-oblongata. I base this opinion, mainly, on a careful consideration of the symptoms, though it will be remembered that in one of Friedreich's cases this organ was found to be the seat of extensive and evidently slowly advancing disease. At the same time I am disposed to think that the cerebellum is also involved in the morbid process, the vertigo exhibited by many of the patients, the nystagmus so constant a feature, the character of the gait being that of a drunken man, and differing entirely from that of ataxia, the pain in the back of the head which existed with several of the patients all point to the presence of cerebellar disease. That the spinal cord is also implicated is beyond question, but the lesion is certainly not limited to the posterior columns, which are not even its chief seat. Indeed, the whole thickness of the cord including the membranes is certainly involved.

If it be objected that in accordance with the observations of Türck, secondary degenerations ascend in the posterior columns and descend in the antero-lateral, I have to cite the experiments of Westphal,[1] which establish the fact that the morbid process extends in the cord in both directions, and along the course of the antero-lateral and posterior columns. Westphal's researches on this point have been amply confirmed by Vulpian,[2] who had at first doubted his conclusions.

I think, therefore, that the affection under notice must be regarded as a distinct form of disease, probably originating in the cerebellum and medulla-oblongata and gradually extending to the spiral cord, which latter is therefore its secondary seat.

---

[1] *Virhow's Archiv* B. xlviii, p. 516, and *Archiv für Psychiatrie.* B. ii.

[2] *Archives de Physiologie,* t. iii, p. 4.